Thorax 1986;**41**:217–218

Short reports

Interrupted aortic arch associated with Pourfour du Petit syndrome

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The anatomy and physiology of the cervical sympathetic chain is well known. This structure is important in the regulation of facial temperature and sweating and the size of the palpebral fissure and pupil.[1] The functions of the cervical sympathetic chain were initially described by Pourfour du Petit. In examining many wartime neck injuries he had the opportunity to observe injuries to the cervical sympathetic chain.[2] He noted signs of increased sympathetic activity in the face and related these to injuries of the cervical sympathetic chain. The syndrome of sympathetic irritative hyperfunction is known as Pourfour du Petit syndrome but has rarely been reported, though the opposite syndrome of cervical sympathetic paralysis is well known as Horner's syndrome. We present a case report of a patient with type B interruption of the aortic arch and associated cervical sympathetic hyperfunction. To the best of our knowledge, this association has not previously been described.

Case report

An 8 year old boy from Colombia was referred with a history of frequent headaches and a heart murmur. On physical examination there was appreciable facial asymmetry, with prominence of the right side of the face and differing skin temperature and sweating. The skin of the right side of the face was cool and showed considerable diaphoresis, even at rest; the left side appeared to be of normal temperature and moisture. The right pupil was 5 mm in diameter and the left 3 mm, dilating normally in the dark. He had considerable proptosis of the right eye (fig 1). There was prominent pulsation over the region of the right carotid artery and his head bounced with a cardiac impulse. A palpable thrill was felt over the right carotid artery. His precordium was hyperactive and the point of maximal impulse was displaced to the left of the midclavicular line, in the fifth intercostal space. There was a prominent thrill in the suprasternal notch and a grade IV/VI harsh systolic murmur, loudest at the left upper sternal border, with radiation to the carotid arteries.

All pulses in the extremities were appreciably diminished. Blood pressure, recorded as 90 mm Hg systolic, could only be obtained by doppler. A chest radiograph showed left ventricular hypertrophy and right apical opacity.

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Accepted 8 July 1985

At cardiac catheterisation, a type B interrupted aortic arch was identified. The right carotid artery arose as a first branch of the ascending aorta and the left carotid artery arose separately as the only other branch of the ascending aorta. The left subclavian artery arose from the descending aorta as did an aberrant right subclavian artery, where the flow was predominantly towards the descending aorta. There were no associated patent ductus or intracardiac anomalies. A magnetic resonance imaging study confirmed an interrupted arch with the ascending aorta giving rise to a considerably dilated and tortuous right common carotid artery (fig 2). There were multiple enlarged vascular structures connected to the aberrant right subclavian artery, which entered the descending aorta below the level of atresia. The patient was taken to the operating theatre and a 12 mm Dacron tube was ana-stomosed end to side with the take off of the left carotid artery proximally and with the descending aorta distally. After operation the patient did well, with marked resolution of his headaches. Though the facial asymmetry persisted there was obvious improvement, the facial skin temperature becoming almost normal and the diaphoresis almost disappearing, and the diameter of the right pupil decreasing to 4 mm. All peripheral pulses were excellent.

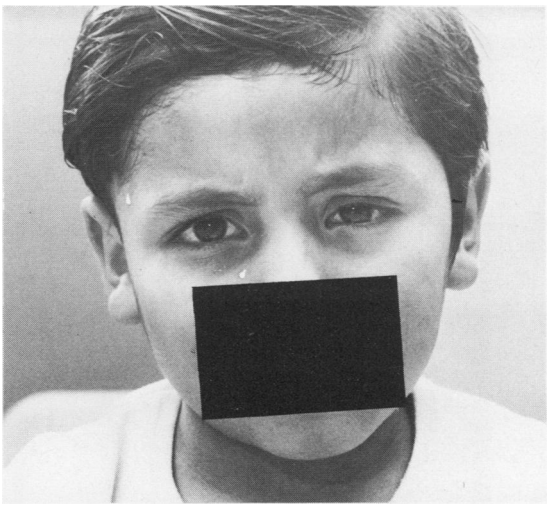

Fig 1 *Photograph of the patient showing diaphoresis and greater size of pupil and palpebral fissure on the right.*

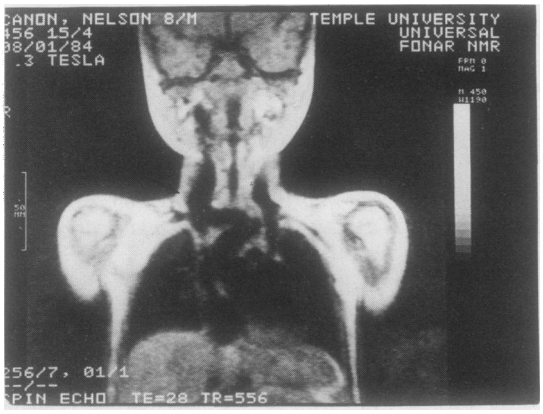

Fig 2 *Nuclear magnetic resonance image showing type B interruption of the aortic arch, a dilated and tortuous right common carotid artery, and a persistent left superior vena cava.*

Discussion

Any compression, irritation, or injury to the sympathetic chain may cause signs and symptoms of sympathetic dysfunction. Though the syndrome of Pourfour du Petit was initially described during the Napoleonic era there have been few cases reported subsequently. In reviewing publications of the last 16 years we were able to find only two documented cases.[3 4] In the case discussed here the classical signs of sympathetic dysfunction were (1) cold skin; (2) increased sweating; (3) mydriasis; (4) widening of the palpebral fissure and proptosis. These signs are exactly opposite to those that would be expected in cases of sympathetic paralysis or Horner's syndrome. In fact, the initial impression when we saw our patient was that a left Horner's syndrome was present. The normal left pupil dilatation in the dark, the clinically obvious increased sweating on the right, and the proved presence of the carotid abnormality close to the right cervical sympathetic chain allowed identification of the exact pathological condition—low cervical sympathetic chain hyperactivity on the right side, presumably secondary to compression and irritation by an enormous, almost aneurysmal, carotid artery. The postoperative improvement probably resulted from a decrease in the tension of the wall of this right carotid artery, secondary to diversion of a large part of the cardiac output through the tube graft from the left carotid artery to the descending aorta.

Possibly increasing compression of the low cervical sympathetic chain may cause sympathetic hyperfunction, which may precede a final phase of sympathetic paralysis described as Horner's syndrome. Awareness of the Pourfour du Petit syndrome may help in the detection of sympathetic dysfunction, and the condition causing it, at an earlier stage and lead to immediate treatment, with improvement of the prognosis.

References

1 Best AE. Pourfour du Petit's experiments on the origin of the sympathetic nerve. *Medical History* 1969;**13**:154–74.
2 Jablonski S. *Illustrated dictionary of eponymic syndromes and diseases and synonyms.* Philadelphia: WB Saunders, 1969.
3 Nappi G, Poloni M, Bono G, Mazzella G, Bo P. Pourfour du Petit syndrome and intracranial aneurysms. *Riv Patol Nerv Ment* 1975;**96**:354–62.
4 Teeple E, Ferrer EB, Ghia JN, Pallares V. Pourfour du Petit syndrome—hypersympathetic dysfunctional state following a direct non-penetrating injury to the cervical sympathetic chain and brachial plexus. *Anesthesiology* 1981;**55**:591–2.